

Acquired pulmonary stenosis due to compression by a bronchiogenic cyst

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A case of pulmonary stenosis due to extrinsic compression of the pulmonary artery and right ventricular infundibulum is reported. It is believed that this is the first such case recorded in the English literature where the cause of the compression was a bronchiogenic cyst.

Pulmonary stenosis due to compression of the right ventricular outflow tract by an extrinsic mass is an uncommon but well-documented event. The recorded causes include lymphoma, teratoma, thymoma, bronchial carcinoma, pericardial sarcoma, and aneurysm of the ascending aorta (Gough, Gold, and Gibson, 1967; Seymour, Emmanuel, and Pattinson, 1968; Littler, Meade, and Hamilton, 1970).

The case reported here appears to be the first in the English literature in which the cause of the compression was a bronchiogenic cyst.

CASE REPORT

The patient was a 3-year-old girl who was referred from Barbados to the University Hospital of the West Indies, Jamaica on 29 October 1970 for investigation of suspected pulmonary valve stenosis. She was free of symptoms but a chest radiograph taken at the time of a respiratory tract infection had revealed a large intrathoracic opacity. The antenatal and obstetric history was normal.

Physical examination showed that she was small for her age but alert and lively. A left-sided praecordial bulge was present and the apex beat was in the left fifth intercostal space on the nipple line. A systolic thrill was palpable along the left sternal border and there was an ejection systolic murmur which was loudest in the third and fourth intercostal spaces just to the left of the sternum. Signs of cardiac failure were absent. There was a dull percussion note and absence of breath sounds over the upper half of the left side of the chest anteriorly. No other abnormal physical signs were found.

INVESTIGATIONS Chest radiographs (Fig. 1) showed a large mass in the position of the left upper lobe. An electrocardiogram revealed gross right ventricular

dominance with a mean QRS axis of $+240^\circ$. Cardiac catheterization was performed and a pressure gradient of 45 mmHg across the pulmonary valve was demonstrated (Fig. 2). Angiocardiograms showed that the infundibulum, the main pulmonary artery, and left pulmonary artery were all pushed inferiorly and to the left by a large, avascular mass in the position of the left upper lobe (Fig. 3).

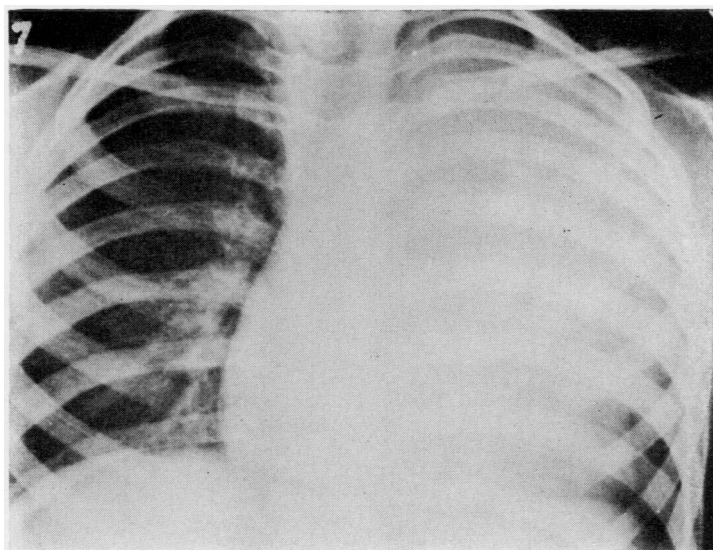
OPERATION A large cystic mass was removed by median sternotomy on 9 November 1970. The mass was situated in the superior anterior mediastinum, extending inferiorly in front of the right ventricle and laterally to fill the upper third of the left hemithorax. It occupied the bay between the ascending aorta and the main pulmonary artery, compressing the latter and the adjacent infundibulum of the right ventricle. The right ventricle was hypertrophied proximal to the site of compression.

The mass was found to be extrapleural and extrapericardial and showed no evidence that it infiltrated adjacent tissues. It was not connected to the trachea or bronchi. After aspiration of 50 ml of thick white mucus, it was removed completely. A withdrawal pressure tracing across the right ventricular outflow tract after removal of the mass showed a residual, but greatly reduced, systolic pressure gradient of 20 mmHg.

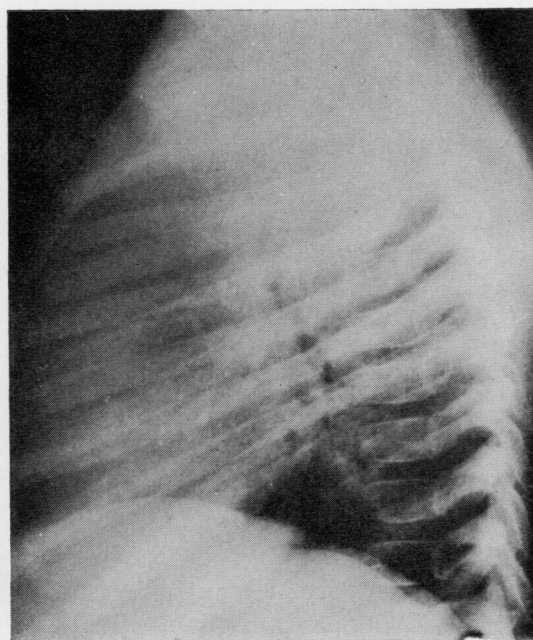
PATHOLOGY The excised specimen consisted of a cystic mass (8 × 5 × 3 cm; weight 70 g) with a smooth, intact outer surface. It was thinly encapsulated and its cut surface showed multiple thin-walled, cystic spaces filled with translucent grey mucin. The lining of the cysts was smooth. In places, small solid zones of grey or white tissue were present between the cysts (Fig. 4).

HISTOLOGY (Fig. 5) Multiple sections were examined and showed that the cysts were lined by ciliated respiratory epithelium which included many goblet cells. Beneath the epithelium were fibrous connective tissue, bundles of smooth muscle, and irregular plates of cartilage. The solid areas described above con-

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(a)



(b)

FIG. 1. *Chest radiographs showing a large anterior mediastinal mass extending into the position of the left upper lobe.*

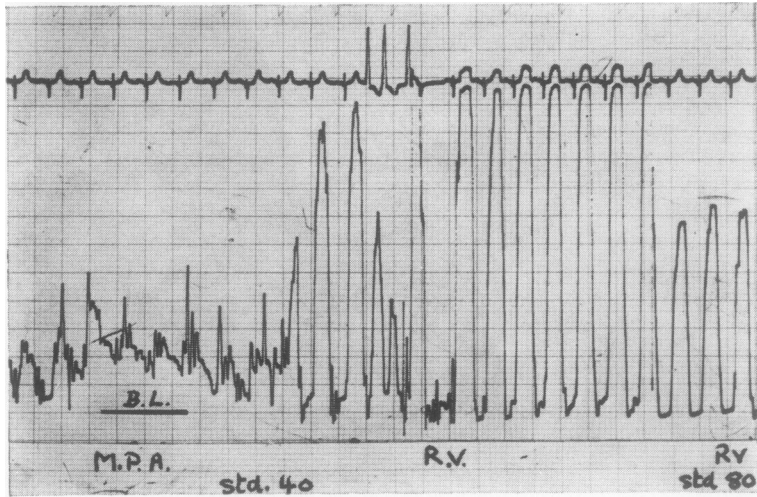


FIG. 2. Cardiac catheterization: withdrawal pressure tracing from the main pulmonary artery (M.P.A.) to the right ventricle (R.V.) showing a gradient of 40 mmHg across the right ventricle outflow tract. The line B.L. indicates the base line.

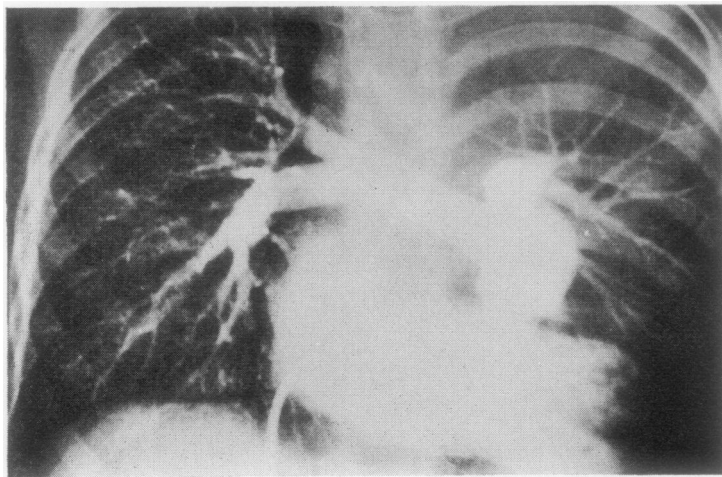


FIG. 3. Angiocardiogram showing displacement and compression of the right ventricular outflow tract by a large mass.

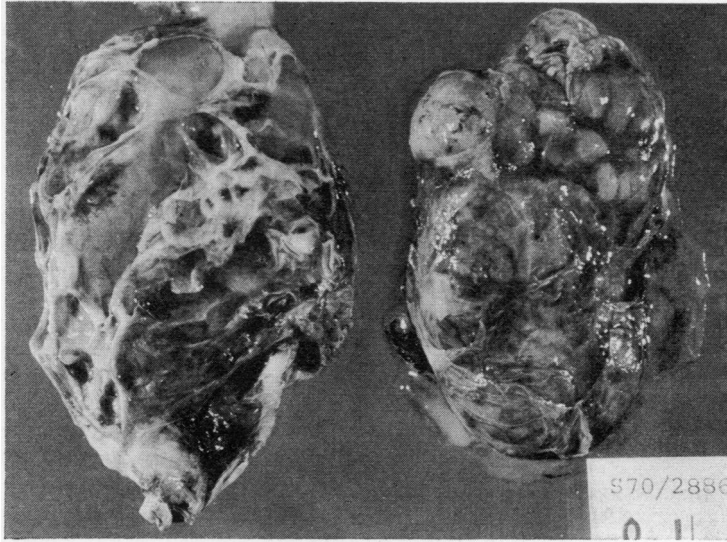


FIG. 4. *The gross pathological specimen. The external surface is seen on the right and the cut surface is on the left.*

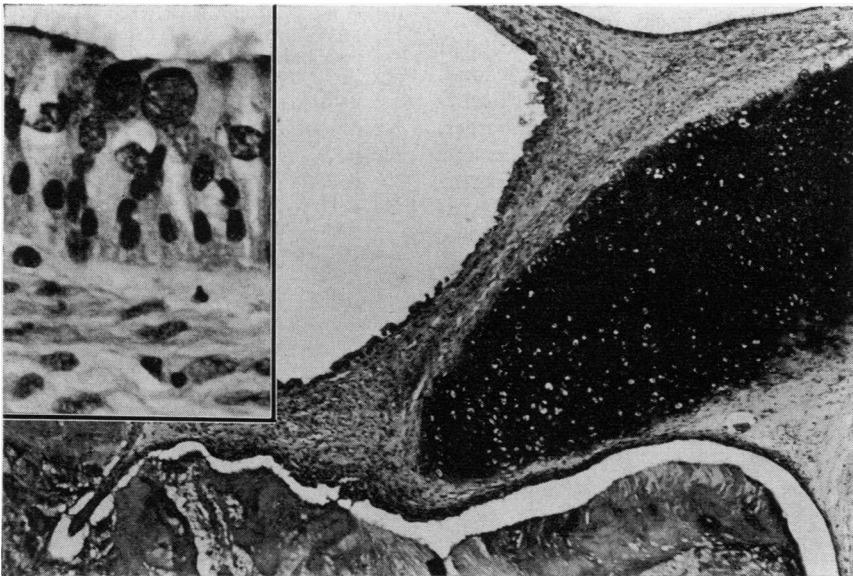


FIG. 5. *Histology of the cyst showing its multiloculated character, epithelial lining, and a plate of cartilage. The lower locule contains mucin. Haematoxylin and eosin $\times 54$. The inset shows the ciliated respiratory-type lining epithelium with goblet cells. Smooth muscle is seen beneath the epithelium. H. and E. $\times 560$.*

sisted of cartilage embedded in fibrous tissue. In some areas the lining epithelium was absent and there was chronic inflammation, apparently associated with extravasation of mucin into the connective tissues. The only tissues found were of the types normally present in the bronchial tree. There was no evidence of neoplasia.

FOLLOW-UP Postoperative convalescence was uneventful. In the immediate postoperative period the pulmonary systolic murmur was still audible but one year later the patient was well and gaining weight and the murmur was no longer present. The disappearance of the residual signs of infundibular obstruction over a period of time is attributable to regression of compensatory infundibular hypertrophy and is similar to the situation following surgical relief of intrinsic pulmonary stenosis with secondary infundibular stenosis (Brock, 1961).

DISCUSSION

The histological appearances indicate that this is a developmental cyst of bronchial origin and we have therefore classified it as a bronchiogenic cyst. This is the commonest form of mediastinal cyst (Abell, 1956) and is thought to arise from the developing lung bud (Fallon, Gordon, and Lendrum, 1954). However, the example described here differs from the usual bronchiogenic cyst in three major respects. First, its multiloculated structure is unusual, the usual form being unilocular; Abell found only three multiloculated examples among 17 cysts of this type. Second, the position of the cyst is uncommon, most occurring in the posterior mediastinum (Schlumberger, 1951); however, this by no means excludes our diagnosis since a variable position within the mediastinum is well recognized, and examples of intrapericardial and even presternal bronchiogenic cysts have been described (Maier, 1948). Third, the occurrence of major complications from such a cyst is uncommon. In Abell's series of 17 cases only five had symptoms due to pressure from the

cyst, and according to Maier such pressure symptoms are usually referable to tracheal or bronchial compression. However, Maier also mentions two cases of intrapericardial bronchiogenic cyst where sudden death was attributed to pressure on the heart or great vessels. The lack of symptoms produced by bronchiogenic cysts doubtless explains the late age at which they are usually discovered (the average age was 36 years in Abell's series) and the present case conforms to the general rule that their discovery is usually accidental. This case appears to be unique in that, although the patient was free from symptoms, the cyst was producing a moderate degree of stenosis of the pulmonary artery. The rarity of this type of complication is attributable to the fact that the usual site of bronchiogenic cysts is in the posterior mediastinum where they do not impinge upon the pulmonary artery.

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